Case Report

Surgical treatment of gastrointestinal hereditary hemorrhagic telangiectasia

Min-Hsuan Yen, and Chiung-Nien Chen

Department of Surgery, National Taiwan University Hospital, Taipei, Taiwan

Case: A 48-year-old man with a history of gastrointestinal bleeding from Osler–Weber–Rendu disease presented with recurrent hematemesis and tarry stool. He received repeated endoscopic therapy, but profound component therapy was still needed. Because repeated gastrointestinal bleeding was caused by same bleeder, tattoo-assisted laparoscopic gastric wedge resection was carried out.

Outcome: The pathology showed vascular abnormalities that involved gastric mucosal and submucosal layers. After surgery, the blood transfusion for the patient is not seen.

Conclusion: Osler–Weber–Rendu is a hereditary disease characterized by vascular abnormalities of the nose, skin, lung, brain, and gastrointestinal tract. Management of gastrointestinal bleeding requires medical treatment first, and there are rare reports of surgical treatment. Our pathology findings showed a transmucosal vessel lesion, which had poor response to endoscopic treatment. Surgical intervention may be considered in the patient with gastrointestinal bleeding refractory to endoscopic therapy.

Key words: Gastrointestinal bleeding, surgery, hereditary hemorrhagic telangiectasia

INTRODUCTION

HEREDITARY HEMORRHAGIC TELANGIECTASIA (HHT), also named Osler–Weber–Rendu disease, is an autosomal dominant disease with an estimated prevalence of 1/5,000.[1] The vascular abnormality is characterized by telangiectasia and arteriovenous malformations in specific locations. Telangiectasia in the nasal and gastrointestinal (GI) mucosa and brain arteriovenous malformations generally present with hemorrhage. Approximately 80% of patients have gastric and small intestinal telangiectasia during examination,[2] but only 25–30% of patients will develop symptomatic GI bleeding until the 5th decade of life.[3,4]

Treatment of GI bleeding includes iron supplementation, component therapy, hormonal therapy,[5] antifibrinolytic therapy, and local endoscopic therapy.[3,6] However, there have been few published reports discussing the role of surgical intervention.[7] No standard surgical approach to recurrent bleeding from HHT lesions has been established. Only one case report has been published regarding the surgical approach to recurrent bleeding from HHT.[8] We present a case in which a preoperative endoscopic tattoo and laparoscopic approach was used to achieve partial gastrectomy for HHT bleeding refractory to endoscopic therapy. We also report the pathology findings.

Corresponding: Chiung-Nien Chen, Professor of Surgery, Department of Surgery, National Taiwan University Hospital, No.7 Chung-Shan South Road, Taipei 100, Taiwan. E-mail address: cnchen@ntu.edu.tw.

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CASE

A 48-YEAR-OLD MAN HAD telangiectasia of the tongue and multiple fingers since the age of 20 years. Intermittent epistaxis was also noted since the age of 26 years. His mother and younger brother also have a history of telangiectasia and epistaxis.

Gastrointestinal bleeding was noted since the age of 45 years, with approximately three episodes in 1 year; the symptoms included tarry stools and hematemesis. Esophagogastroduodenoscopy showed angiodysplasia (Fig. 1A). At first, the bleeding responded well to endoscopic therapy (argon plasma coagulation and hemoclipping). But the symptoms became more severe, and the effect of endoscopic therapy had been limited in the last 3 months. Argon plasma coagulation, sclerotherapy, adrenalin injection, and hemoclipping were applied, but the GI bleeding still progressed. The patient had hematemesis and bloody stool, and his hemoglobin decreased from 12 g/dL to 8.1 g/dL, even after a 12-unit blood transfusion 1 month previously. We found the same bleeder at the posterior wall of gastric middle body leading to active bleeding in recent endoscopic therapy

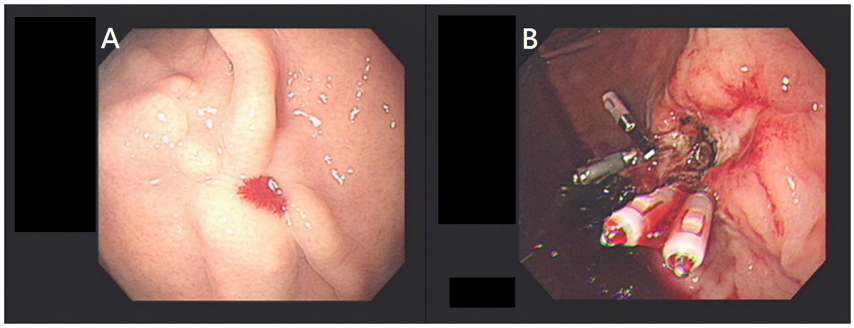

Fig. 1. (A) Angiodysplasia at the posterior wall of gastric middle body of a 48-year-old man with hereditary hemorrhagic telangiectasia. (B) The condition had proved refractory to the several endoscopic treatments.

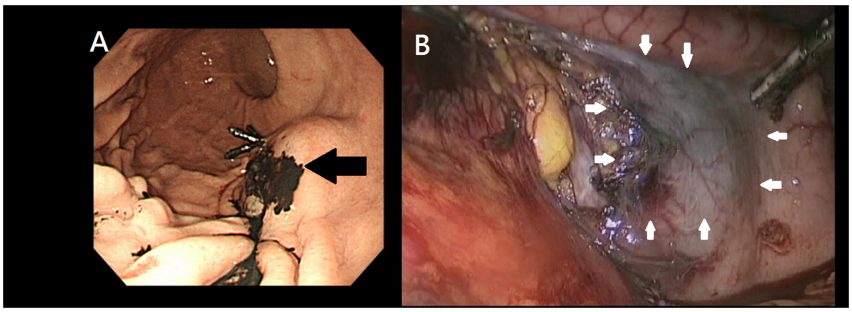

Fig. 2. Tattoo for localization of gastrointestinal bleeding in a 48-year-old man with hereditary hemorrhagic telangiectasia. Black arrows, endoscopic view; white arrows, laparoscopic view.

(Fig. 1B). Computed tomography angiography of the patient showed one 10-mm angiodysplasia at the posterior wall, which was compatible with endoscopic findings. According to our institution's gastroenterologist and radiologist, the larger size indicated the higher rate of rebleeding. After discussing with the patient, surgical intervention was determined to be the most appropriate therapy. We chose the endoscopic tattoo method (Fig. 2A) for pre-operative localization of the main active bleeder, because HHT is characterized by multiple lesions. Laparoscopic and endoscopice cooperative surgery was not chosen, as we were unsure we would be able to localize the bleeding during the operation. When the main active bleeder was identified by panendoscopy, we carried out the laparoscopic wedge resection the next day.

During surgery, an infraumbilical 10-mm port, one right 10-mm port, and one left 5-mm port were created. In the beginning, we noted telangiectasias over the liver surface. After mobilizing the greater curvature, the tattoo site (Fig. 2B, white arrows) was identified at the posterior wall near the lesser curvature. Gastric wedge resection was carried out by five reloads of 60-mm endo linear cutter staplers; specimen examination confirmed complete removal of the arteriovenous malformation lesion.

The pathology (Fig. 3) showed angioectasia of abnormal vascular malformation involving mucosa and submucosa of stomach. Within the submucosa was the vascular malformation, presenting marked tortuous vessels with irregular wall thickness (Fig. 3, black arrows). Tattoo dye (Fig. 3, white arrows) in the submucosal layer was also seen. There was focal acute, fresh, mucosal hemorrhage, consistent with a sequel of repeated gastric bleeding. Its surgical margins were free.

One year after surgery, some episodes of GI bleeding were noted. However, this could be managed by conservative treatment without the need for component therapy. The patient resumed a normal diet on post-operative day 5. There were no digestive GI symptoms after surgery.

DISCUSSION

THE TREATMENT OF GI bleeding in HHT includes component therapy or iron supplements for mild anemia and endoscopic therapy for cases refractory to component therapy. Hormonal therapy or antifibrinolytic therapy is effective in some groups of patients. There are also case reports describing the use of antiangiogenic agents[9] like bevacizumab and thalidomide.[10] However, there are very few published works discussing the use of surgery for GI bleeding in HHT.

According to pathology report, we revealed the involvement of the depth. The most effective treatment for GI bleeding is argon plasma coagulation. The maximum depth of the argon plasma coagulation zone was approximately 3–5 mm.

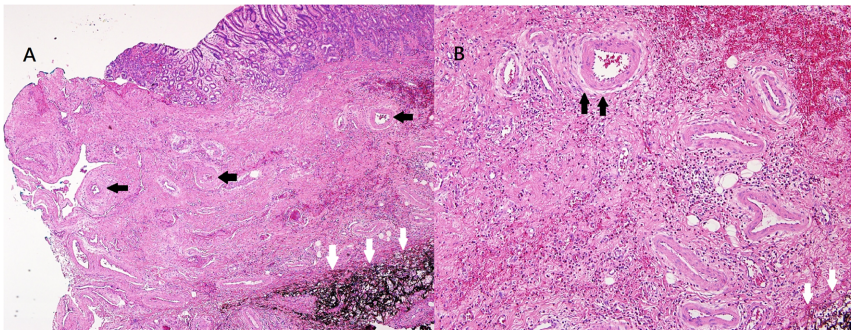

Fig. 3. Vascular malformation in a 48-year-old man with hereditary hemorrhagic telangiectasia. Tortuous vessels with irregular wall thickness (black arrows) and tattoo carbon ink (white arrows) in the submucosal layer are shown at 40× (A) and 100× (B) magnification.

It is difficult to treat the whole range of the lesion. We assume that symptomatic GI bleeding of HHT that is refractory to endoscopic treatment may reserve surgical intervention for this subgroup of arteriovenous malformations. Multiple attempts will just increase the suffering of the patients who already received little relief from this kind of disease. Gastric wedge resection would be more precise using perioperative localization.[8] Endoscopic localization will reduce the damage of this surgery by preserving the gastric residual volume.

CONFLICT OF INTEREST

NONE.

REFERENCES

1 Kjeldsen AD, Vase P, Green A. Hereditary haemorrhagic telangiectasia: A population-based study of prevalence and mortality in Danish patients. J. Intern. Med. 1999; 245: 31–9.
2 Ingrosso M, Sabba C, Pisani A *et al*. Evidence of small-bowel involvement in hereditary hemorrhagic telangiectasia: A capsule-endoscopic study. Endoscopy 2004; 36: 1074–9.
3 Faughnan ME, Palda VA, Garcia-Tsao G *et al*. International guidelines for the diagnosis and management of hereditary haemorrhagic telangiectasia. J. Med. Genet. 2011; 48: 73–87.
4 Kjeldsen AD, Kjeldsen J. Gastrointestinal bleeding in patients with hereditary hemorrhagic telangiectasia. Am. J. Gastroenterol. 2000; 95: 415–8.
5 Marshall JK, Hunt RH. Hormonal therapy for bleeding gastrointestinal mucosal vascular abnormalities: A promising alternative. Eur. J. Gastroenterol. Hepatol. 1997; 9: 521–5.
6 Longacre AV, Gross CP, Gallitelli M, Henderson KJ, White RI Jr, Proctor DD. Diagnosis and management of gastrointestinal bleeding in patients with hereditary hemorrhagic telangiectasia. Am. J. Gastroenterol. 2003; 98: 59–65.
7 McDonald J, Bayrak-Toydemir P, Pyeritz RE. Hereditary hemorrhagic telangiectasia: An overview of diagnosis, management, and pathogenesis. Genet. Med. 2011; 13: 607–16.
8 Park J, Ellis B, Juergens C. Laparoscopic resection of Osler–Weber–Rendu lesion. JSLS 2008; 12: 180–2.
9 Bauditz J, Lochs H. Angiogenesis and vascular malformations: Antiangiogenic drugs for treatment of gastrointestinal bleeding. World J. Gastroenterol. 2007; 13: 5979–84.
10 Wang XY, Chen Y, Du Q. Successful treatment of thalidomide for recurrent bleeding due to gastric angiodysplasia in hereditary hemorrhagic telangiectasia. Eur. Rev. Med. Pharmacol. Sci. 2013; 17: 1114–6.